



North Shore-Long Island Jewish Health System



HARVEY  
CUSHING  
institutes of  
neuroscience

## New Patient Visit

Name of the patient: [REDACTED]

DOB: [REDACTED]

Visit date: 3/25/08

PCP: Rhonda Westcott, MD  
(208) 292-5437

Ref Phys:

### Report distribution:

PCP: ☒ Ref Phys: ☐ Patient: ☒ Other: ☐

### Chief complaints today are:

1. Headache
2. Temporary Loss of Vision and Color Change
3. Vomiting

### Associated brainstem symptoms:

- vomiting, snoring

Lansky score today is:

80-90

Beighton score today is:

7

Pain assessment (1-10 scale):

0/10 as per mom

### History of present illness:

[REDACTED] is a [REDACTED] old little girl who presents to The Chiari Institute today accompanied by her parents, from Hayden, Idaho. [REDACTED] was recently diagnosed with Chiari I Malformation, and is here for further evaluation and treatment options.

[REDACTED] was born at 36 weeks gestation, a twin, conceived via IVF. [REDACTED] parents report that she experienced reflux and constipation in her first year of life. She was also diagnosed with a speech delay, but otherwise met her developmental milestones on time.

On January 16<sup>th</sup>, 2008, [REDACTED] was jumping in the playground, when she began to complain of a severe headache and loss of vision. She was brought to an urgent-care office, diagnosed with a migraine, and sent home.

Her mother reports that she continued to have nausea and vomiting, and was also experiencing visual color identification difficulties. At this time, she also had difficulty with dizziness and imbalance. Her parents brought her to the Emergency Department, where an MRI was

performed, and she was diagnosed with Chiari I Malformation. Dr. Ling, a pediatric neurosurgeon, was consulted, and reportedly recommended immediate surgery.

symptoms lasted approximately one week, and then resolved spontaneously. A repeat episode, similar to the first, occurred about 2 weeks ago, lasting about 3 days.

parents then performed an internet search, found The Chiari Institute, and have brought her here today.

**Headache Characterization:**

Every once in a while c/o headache, usually suboccipital headache

**Location:** suboccipital ☒ frontal ☐ temporal ☐ vertex ☐

**Radiation:** cervical ☐ retro-orbital ☐ frontal ☐ temporal ☐

**Sidedness:** right ☐ left ☐ bilateral ☐

**Quality:** sharp ☐ pounding ☐ pressure ☐ dull ache ☐ electric-like ☐  
burning ☐

**Intensity:** without medication 4-5/10 with medication 0/10

**Symptom Associations:** nausea ☐ vomiting ☐ scotoma ☐ scintillations ☐  
photophobia ☒ phonophobia ☒

**Temporal associations:** daily ☐ menstrual ☐ nocturnal with awaking ☐  
present on awaking ☐ end of day ☐

**Duration:** fleeting ☐ hours ☐ days ☐ continuous ☐ comes & goes ☐

**Relieving factors:** usually Tylenol and nap help headache  
medication ☒ lying down ☒ wearing neck collar ☐

**Aggravating factors:** cough ☐ laughing ☐ singing ☐ crying ☐  
sneezing ☐ lifting ☐ straining at stool ☐ exercise ☐  
orgasm ☐  
neck: flexion ☐ extension ☐ rotation ☐

**Past lumbar puncture findings:**

Denies

**Past trauma history:**

Denies

**Past surgical history:**

Denies

**Past medical history:**

1. constipation

2. reflux

**Review of Systems:**

**Denials:**

- palpitations,
- syncope, lightheadedness,
- dizziness, vertigo;
- dysphagia,
- nausea, vomiting,
- constipation, diarrhea;
- urinary urgency, hesitancy or incontinence, enuresis;
- excessive sweating;
- reversible joint dislocations, congenital joint dislocations, or hyperextensible joints.
- insomnia, early morning arousal;
- sleep position (stomach, side, back); all over
- excessive daytime sleepiness;
- snoring – irregular/rhythmic;
- chronic fatigue
- position of legs during sleep: all over
- depression, bipolar disorder, anxiety/panic disorder, thought disorder, paranoia, and hallucinations;
- diabetes, thyroid disease, coronary artery disease, irritable bowel syndrome, asthma, sleep apnea, history of Lyme disease;
- problems thinking, feeling as if she is in a “fog,”
- problems with language – production or comprehension

**Medications (list all including OTC drugs and supplements):**

Fluoride 0.5mg qd  
Tylenol prn (few times/month)

**Allergies:**

Patient denies allergies to any medication, foods, iodinated dye, gadolinium or latex.

**Family history:**

Maternal: GF, 47 Chronic neck pain  
GM, 47 healthy  
Paternal: GF 52, arthritis  
GM 51, DM, HTN, high Cholesterol  
Mother 27, CMI, polycystic ovaries, In vitro fertilization  
Father 30, healthy  
Twin brother, healthy

**Developmental History:**

She was born at 36 ½ weeks as a twin through In Vitro fertilization, 5lbs, 5 oz, NSVD.

Pt had reflux for 2 years, some changes in her voice.

Language is average but speech delayed, receiving speech therapy.

**Social history:**

Education: pre-K

Tobacco: NA

Alcohol: NA

Caffeine: NA

Recreational Drugs: NA

**Physical Exam:**

The patient is in no apparent distress.

Handedness: right

BP: 90/60 HR: 96 and regular Wt: 32 lbs.

HC: 51.5 cm (normocephalic)

The chest is clear; abdomen soft and nontender.

There is no peripheral edema, clubbing or cyanosis.

No scoliosis or cutaneous stigmata are noted.

Joint hypermobility is noted (see Beighton score); TMJs are intact

No limb length discrepancies are noted.

**Neurological Exam:**

Pt. is alert, playful and appropriately oriented.

She is cooperative; mood and affect are normal.

Speech output is somewhat decreased; no dysphasia is noted.

Fundoscopic exam is benign; discs flat, no venous pulsations are seen.

Visual fields are full to confrontation.

EOMs are intact, PERRL, no nystagmus, no ptosis.

No facial asymmetry.

Hearing is intact to finger rub. EAC in normal.

Tongue, palate and pharynx intact; gag reflex is present bilaterally.

No dysarthria or dysphonia is noted.

Walking and running are normal; some difficulty in hopping with right leg; heel and toe walking are normal.

No difficulty with tandem standing and walking is noted.

Muscle strength and bulk are intact; no drift is noted.

DTRs 1-2+ and symmetrical; plantar responses are flexor.

Coordination and rapid alternating movements are intact.

Tone is normal; no abnormal movements or tremors are noted.

Romberg (Std) negative.

Sensation is grossly intact.

**ROM Assessment:**

Neck: rot R(80°): 80° rot L(80°): 80° Ext(50°): 50° Flex(60°): 60°

**Provocative tests:****Cranio-cervical Junction:**

- |                           |               |
|---------------------------|---------------|
| 1. Axial loading:         | Some headache |
| 2. Extraction (sitting):  | No change     |
| 3. Extraction (standing): | No change     |
| 4. Jugular compression:   | No change     |

**Lumbar Spine**

- |                          |           |
|--------------------------|-----------|
| 1. Heel walking:         | Negative  |
| 2. Toe walking:          | Negative  |
| 3. Pelvic traction:      | Back pain |
| 4. Straight leg raising: | Negative  |
| 5. ROM at hips:          | Normal    |

**Neuroradiological studies:**

- Tonsillar herniation extending between C1 and C2, pegged shaped
- Cerebellar prolapse
- Impaction of CSF spaces and flow at the CCJ
- Distal slit-like central syrinx, along the last segments of the Thoracic spinal cord
- Conus ending at L1-2
- The proximal part of the Filum Terminale is thick and remains so up to L2-3
- Straight L spine
- No lumbarization of S1

Occipital Bone	small
Posterior cranial fossa	small
Brain stem	elongation
Downward displacement of brain stem	short clivus

**Assessment:**

The patient is affected by the following pathologies:

- Chiari I Malformation (CMI)
- Ehlers-Danlos Syndrome (EDS)
- No evidence of Functional Cranial Settling (FCS)
- Tethered Cord Syndrome (TCS).

The tonsillar herniation is the sum of the effects of the CMI and the TCS.

The form of TCS which is currently suspected is the Occult variant.

Morphometric Measurements detected the following:

- The volume of the Posterior Fossa is small, and c/w the diagnosis of CMI

- Elongation and downward displacement of the brainstem, which in our experience are quite specific signs of Tethered Cord Syndrome (TCS).

Lansky score today is: 80-90  
Beighton score today is: 7

**Plan:**

In our opinion, the patient is a candidate for the surgical management of her TCS, as follows:

- Section of the Filum Terminale (SFT) to address the TCS, under Color Doppler and Neuromonitoring guidance
- An MRI of the Brain will be obtained at the time of the hospital discharge
- Three to six months later, the patient will be re-evaluated at TCI, with an MRI of the Brain, Cervical, and Lumbar spine, and a follow-up visit, to assess the residual impact of the tonsillar herniation
- A plastic surgeon should be readied locally, to treat eventual wound healing problems related to EDS.

The risks, benefits, potential complication, and non-surgical alternatives were exhaustively explained to the patient.

In the face of the elevated Karnofsky score, the patient is a candidate for conservative management of her CMI. As part of the conservative management she will:

- Be treated symptomatically;
- Practice risk avoidance (whiplash injuries and strenuous physical straining);
- Be clinically monitored, with yearly neuroimaging and follow-up evaluations at TCI.

It was a pleasure to see Ms [REDACTED] at the Chiari Institute. It is also a pleasure to pleasure to participate with you in her care.



Paolo Bolognese, M.D. (Neurosurgery)  
Associate Director, The Chiari Institute  
Harvey Cushing – Institutes of Neuroscience  
North Shore – LIJ Health System

## Ehlers-Danlos Screening

### Beighton score

Thumb to forearm (1 point for each arm for possible total of 2)	<input checked="" type="checkbox"/> R <input checked="" type="checkbox"/> L
Bending little fingers back to 90° or more (1 point for each hand for a total of 2 points)	<input checked="" type="checkbox"/> R <input checked="" type="checkbox"/> L
Elbows hyperextend to 10° or more (i.e. bend backwards 10° or more in the wrong direction) (1 Point for each arm for a total of 2 points)	<input type="checkbox"/> R <input checked="" type="checkbox"/> L
Knees hyperextend to 10° or more (i.e. bend backwards 10° or more in the wrong direction) (1 point for each leg for a total of 2 points)	<input checked="" type="checkbox"/> R <input type="checkbox"/> L
Palms flat to floor with legs straight (1 point)	<input checked="" type="checkbox"/> Able <input type="checkbox"/> By history
TOTAL SCORE	7

For a total of 9 points, a score of 4 or more is indicative of hypermobility.

Wound healing problems	no
Spontaneous joint dislocation	no
Congenital hip dislocation	no
Vascular type features (translucent skin, arterial/intestinal/uterine fragility or rupture, tendon and muscle rupture can occur. Talipes equinovarus (clubfoot) is frequently seen at birth. Other manifestations that may be found in the Vascular Type include: acrogeria (premature aging of the skin of the hands and feet); early onset varicose veins; arteriovenous fistula (an opening between an artery and vein), carotid-cavernous fistula; pneumothorax (collapse of a lung) /pneumohemothorax (collapse of a lung with a collection of air or gas and blood); gingival recession and complications during and after surgery (i.e. wound dehiscence).	no
MVP	no

wk





North Shore-Long Island Jewish Health System

The Chiari Institute

## Tethered Cord Syndrome Screening Tool

Please answer the questions by typing X or ✓ in the appropriate column

as per mom

Questions	Yes	No
Do you have urinary urgency? <u>sometimes</u>	<input checked="" type="checkbox"/>	
Do you urinate often? How many times? <u>15</u> <u>sometimes</u>	<input checked="" type="checkbox"/>	
Do you have urinary incontinence? Dribbling? _____ Drenching? _____ <u>Potty trained; wears pull ups night</u>	<input checked="" type="checkbox"/>	
Do you have difficulty emptying your bladder completely?		<input checked="" type="checkbox"/>
Do you urinate at night? How many times? _____ <u>pulls ups at night</u>	<input checked="" type="checkbox"/>	
Do you have problems starting your urinary stream?		<input checked="" type="checkbox"/>
Do you have constipation?	<input checked="" type="checkbox"/>	
Do you have diarrhea?		<input checked="" type="checkbox"/>
Do you have occasional incontinence for stools?		<input checked="" type="checkbox"/>
Do you have decreased interest in sexual relations?		na
Do you have difficulty reaching an orgasm?		na
Do you have decreased sensation in your pelvic area?		na
Do you have low back pain?		<input checked="" type="checkbox"/>
Do you have leg pain?		<input checked="" type="checkbox"/>
Do you have restless leg syndrome?		<input checked="" type="checkbox"/>
Do you have numbness under the soles of your feet?	<input checked="" type="checkbox"/>	
Do you keep your knees bent at night to relieve back or leg discomfort?		<input checked="" type="checkbox"/>
Do you have low back pain, leg pain, or urinary symptoms while walking up stairs? <u>In am only</u>	<input checked="" type="checkbox"/>	
Do you have a history of severe growing pains during childhood and adolescence?		na
Have you had urological surgery? If yes, when? _____		<input checked="" type="checkbox"/>
Have you had urodynamic testing? If yes, when? _____		<input checked="" type="checkbox"/>

wk

Filum Terminale Traction Tests	Positive	Observed symptoms
Heel walking	No	
Toe walking	No	
Pelvic traction	<input checked="" type="checkbox"/> Yes	LBP

Questions	Yes	No
Has a terminal syrinx been identified?	<input checked="" type="checkbox"/>	
Has scoliosis been identified?		<input checked="" type="checkbox"/>



# Morphometrics

Occipital Bone		
Structure	Length (mm)	Normals Mean ( $\pm$ SD)
Clivus	34.3	47.2 (45-49)
Supraocciput	41.9	47.7 (45-50)
Condyle	18.8	24.1 (22-26)

Date of the imaging studies from which these measurements were obtained:

██████████  
All county, 3/24/08

Brain					
Structure	Length (mm)	Normals Mean ( $\pm$ SD)	Angle	Degrees (°)	Normals Mean ( $\pm$ SD)
Brainstem Length	55.9	50.5 (48-53)	Tentorial Supraoc. Angl.	77	83.4 (82-88)
Ponto-medullary Ht	35.5	42.4 (39-47)	Twining Tent. Angl.	40	37.9 (34-41)
Tentorial Length	44.8	48.1 (45-51)	Basal Angl.	123	124 (117-132)
Medullary Ht	14.0	18.5 (14-23)			
Tonsillar Descent	17.9				

Cranio-Cervical Junction				
Structure	Supine		Upright	
	Length (mm)	Normals Mean ( $\pm$ SD)	Length (mm)	Normals Mean ( $\pm$ SD)
Basion-Dens Intrv.		7.4 (5.8-9.0)		7.2 (5.6-8.8)
Basion-Atlas Intrv.		1.8 (0-3.0)		2.1 (0-4.0)
Angle	Degrees (°)		Degrees (°)	
Clival-Axis Angl.		148 (141-154)		147.8 (142-154)
Clival-Atlas Angl.		38.0 (30-46)		36.5 (29-44)

Volume Calculations		
Structure	Volume (ml)	Normals Mean ( $\pm$ SD)
Posterior Cranial Fossa	136.7	188.5 (178.5-198.5)
Posterior Fossa Brain	94.9	151.7 (148.3-155.1)

Impression	
Occipital Bone	small
Posterior cranial fossa	small
Brain stem	elongation
Downward displacement of brain stem	short clivus
Cranial settling	N/A

John X. Chen, M.D.

Paolo Bolognese, M.D.  
Weon Hi Kang, N.P.

9 of 9 pages  
Form Rev. 3/11/08